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IMAGES IN NEONATAL MEDICINE.....

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Prenatal three dimensional ultrasound detection of linear nevus sebaceous syndrome

A full term, male neonate was born to a 38 year old mother by uncomplicated vaginal delivery. Amniocentesis had been performed, and a chromosome study revealed a normal male karyotype at 11 weeks gestation. A prenatal three dimensional sonogram showed two skin tag-like lesions over the left orbital region at 28 weeks gestation (fig 1).

Physical examination of the neonate found linear, well demarcated, yellowish, hairless plaques located on the forehead, scalp, and facial region. Two nodules over the lateral canthus and upper eyelid of the left eye resulted in ectropion (fig 2) There were no other cutaneous lesions. Magnetic resonance imaging of the brain showed hemimegalencephaly. The baby suffered a seizure attack about two weeks after birth.

Linear nevus sebaceous syndrome is a rare sporadic oculoneurocutaneous disorder, consisting of a spectrum of abnormalities involving the skin, central nervous system, eyes, and other systems. The major clinical manifestations include linear nevus sebaceous, seizure, and mental retardation. The aetiology of this disorder is still not identified.¹ The major finding on brain image is hemimegalencephaly, which is characterised by congenital overgrowth of one cerebral hemisphere ipsilateral to the skin lesions. The affected brain has essentially no function, and is often associated with hemiparesis, early onset seizures, mental retardation, hemimacrocephaly, and severe encephalopathy clinically.^{1–3} Prenatal diagnosis is difficult but a fetal sonogram, especially a three dimensional image, may be useful for early detection of linear nevus sebaceous syndrome.^{4–5}

S-H Lien, M-L Hsu, Y-S Yuh, C-M Lee

Department of Pediatrics, Tri-Service General Hospital, Taipei, Taiwan, ROC



Figure 1 Skin tag-like lesions over left orbital region seen on three dimensional image at 28 weeks gestation.

C-C Chen, P-Y Chang
Department of Pediatrics, Armed Forces Sung Shan Hospital, Taipei, Taiwan, ROC
C-Y Chou
Department of Obstetrics & Gynecology, Armed Forces Sung Shan Hospital

Correspondence to: Dr Lee, Department of Pediatrics, Tri-Service General Hospital, No 325, Cheng-Kung Road, Sec 2, Neihu 114, Taipei, Taiwan, ROC; lcmpin@yahoo.com.tw

Parental consent was obtained for publication of figure 2

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Figure 2 Linear, well demarcated, slightly raised, yellowish, hairless plaques located on the left side of the forehead, the parietal region of the scalp, face, and chin. Congenital ectropion caused by two nodules over the lateral canthus and upper eyelid can be seen.